Ocular masquerade syndrome

Keywords: masquerade, syndrome, pseudo-uveitis, ocular, inflammation, neoplasia, tumor, resistance to steroids

Abbreviations: OMS, ocular masquerade syndrome; JXG, juvenile xanthogranuloma; CAR, carcinoma-associated retinopathy; MAR, melanoma-associated retinopathy; PRL, primary retinal lymphoma; CNS, central nervous system; PCNSL, primary CNS lymphoma.

Editorial

In the ophthalmologic literature, the term “masquerade syndrome” has been firstly used in 1967 for the description of a conjunctival carcinoma presenting as chronic conjunctivitis, an extracocular inflammatory disorder. However, especially a group of malignant and non-malignant systemic or primary ocular diseases mimicking immune-mediated uveitis have been also included into the spectrum of this syndrome. Ocular Masquerade Syndrome (OMS) is often misdiagnosed as chronic uveitis, in the other words, OMS is a group disease of the causes of pseudo-uveitis. However, in OMS, cells arise from a non-inflammatory origin (intraocular pigment, blood, or neoplastic cells). The most common features of this entity are bilateral asymmetrical involvement, the presence of aqueous and/or vitreous cells, older age, history of systemic or ocular malignancy, the absence of other inflammatory signs such as ocular pain, keratic precipitates and synchiae, and initial response and eventual resistance to corticosteroids. OMS usually points out malignant entities including sebaceous gland carcinoma (meibomian gland carcinoma or sebaceous adenocarcinoma), intraocular and periorcular lymphoma, retinoblastoma, diffuse amelanotic ocular melanoma and metastatic tumors to the choroid and retina. The incidence of malignant entities in OMS is 2-8% of cases in a uveitis practice.

However, some non-malignant or non-neoplastic proliferative conditions can be considered OMS. These include uveal juvenile xanthogranuloma (JXG), fungal or chronic bacterial endophthalmitis (P. Acnes infection, etc), scleritis/episcleritis, intraocular foreign body, siderosis, amyl再度is, pigment dispersion syndrome, old vitreous hemorrhage, retinal vascular occlusions or retinal ischemia, irido-corneo-endothelial syndrome (ICE), retinitis pigmentosa, anterior segment ischemia caused by carotid artery disease or irradiation, ocular ischemic syndrome, and chronic peripheral retinal detachment, paraneoplastic syndromes such as carcinoma-associated retinopathy (CAR)/melanoma-associated retinopathy (MAR), bilateral diffuse uveal melanocytic proliferation caused by intraocular dispersion or migration of cells. The most frequent neoplastic cause is primary intraocular or retinal lymphoma (PRL) having a high-grade neoplasia, an aggressive course, neural retinal infiltration, and central nervous system (CNS) involvement. However, primary CNS lymphoma (PCNSL) that is extranodal, non-Hodgkin’s B cell lymphoma involving CNS or eye or both, primary choroidal or iridal lymphoma or secondary intraocular infiltration of a systemic lymphoma may mimic an intraocular inflammatory condition. In the suspicion of a steroid resistant bilateral uveitis in especially patients between 50 and 60 years of age, PRL should be excluded. The most common symptoms of the PRL include painless visual loss and “floaters”. The most common signs are anterior chamber involvement (over 30%), multiple yellow-orange subretinal pigment epithelial lesions (leopard skin appearance on fluorescein angiography).

In contrast, primary choroidal lymphoma (PCL), B cell lymphoma of mucosa-associated lymphoid tissue (MALT) type has a low-grade and a good prognosis. The majority of primary iridal lymphomas have high-grade neoplasia, but relatively good prognosis. The main causes of OMS in adults are PRL, primary intraocular lymphoma, ocular metastasis of systemic Non-Hodgkin lymphoma, or breast/lung/renal carcinomas, or uveal melanomas. In children, OMS usually occurs due to leukemia, retinoblastoma, medulloepithelioma or juvenile xanthogranuloma or persistent hyperplastic primary vitreous. In malignant causes, isolated choriretinal lesions, serious retinal detachment, and iris infiltration can seem while as in nonmalignant conditions, chorioretinal lesions with surrounding cellular infiltrate and pseudo-retinal vasculitis may be observed. The main causes of OMS in adults are PRL, primary intraocular lymphoma, ocular metastasis of systemic Non-Hodgkin lymphoma, or breast/lung/renal carcinomas, or uveal melanomas. In children, OMS usually occurs due to leukemia, retinoblastoma, medulloepithelioma or juvenile xanthogranuloma or persistent hyperplastic primary vitreous. In malignant causes, isolated choriretinal lesions, serious retinal detachment, and iris infiltration can seem while as in nonmalignant conditions, chorioretinal lesions with surrounding cellular infiltrate and pseudo-retinal vasculitis may be observed. The main causes of OMS in adults are PRL, primary intraocular lymphoma, ocular metastasis of systemic Non-Hodgkin lymphoma, or breast/lung/renal carcinomas, or uveal melanomas. In children, OMS usually occurs due to leukemia, retinoblastoma, medulloepithelioma or juvenile xanthogranuloma or persistent hyperplastic primary vitreous. In malignant causes, isolated choriretinal lesions, serious retinal detachment, and iris infiltration can seem while as in nonmalignant conditions, chorioretinal lesions with surrounding cellular infiltrate and pseudo-retinal vasculitis may be observed.

Acknowledgments
None.

Conflict-of-interest
The author declares that there is no conflict of interest regarding the publication of this paper.

Authorship contributions
Concept and design, data collection, literature Search, writing: analysis, interpretation: burak turgut.

References


